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Dissociation between intact vibratory sensation and impaired joint position sensation may predict ataxia of spinal origin

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ABSTRACT

Vibratory and joint position sensations are usually impaired simultaneously and afferents for both sensations ascend the dorsal columns. There are a few evidences that the central pathways in the spinal cord for position and vibratory sensations are not identical. In this study, we examined the clinical features of patients with sensory impairments of vibratory and joint position sensations. According to 43 evaluated patients' results, the dissociation between an intact vibratory sensation and impaired joint position sensation may be important for the diagnosis of spinal disorders. We also report three cases of patients with spinal ataxia caused by sensory impairments and who show the dissociation between an impaired joint position sensation and an intact vibratory sensation. The combination of intact vibration sensation and impaired joint position sensation may suggest a dorsal column lesion in the spinal cord.

1. Introduction

Vibratory and joint position sensations are usually impaired simultaneously and afferents for both sensations ascend the dorsal columns [1]. However, some patients showed dissociation between vibratory and joint position sensation. There are a few evidences that the central pathways in the spinal cord for position and vibratory sensations are not identical [2,3]. In this study, we examined the clinical features of patients with sensory impairments of vibratory and joint position sensations. Herein, we also report three cases of patients with spinal ataxia caused by sensory impairments, who showed the dissociation between an impaired joint position sensation and an intact vibratory sensation. The dissociation between an intact vibratory sensation and impaired joint position sensation may be important for the diagnosis of spinal disorders.

2. Case reports

We performed a retrospective analysis using medical charts to determine the localization, cause, clinical features among consecutive patients with impaired vibratory and/or joint position sensations, who were admitted to our hospital between April 2012 and April 2016. The localization of anatomical lesions was assessed by nerve conduction studies, somatosensory-evoked potentials and/or magnetic resonance images (MRIs) as peripheral nerve, nerve root/dorsal root ganglion, spinal cord, brainstem, or cerebral lesion. The symptom of nerve root/dorsal root ganglion was defined by lesions restricted predominantly to the spinal roots or the very proximal portion of the spinal nerves. The identification of nerve root/dorsal root ganglion lesion was due to presence of F-wave abnormalities, poor occurrence and prolonged latencies of somatosensory-evoked potentials [4]. Patients with diabetes mellitus (HbA1c > 6.5%) or treated with antidiabetic agents were excluded from this study. Vibratory sensation was evaluated using a tuning fork which measures the amplitude threshold of 128 Hz vibration. The definition of impaired vibratory sensation was present if the examiner senses the vibration on patients for <8 s. Joint position sensation was evaluated on the basis of the ability to identify flexion and extension of fingers or toes at different angular velocities, identification of fingers and toes held by an examiner, and changes in titubation of the trunk after eyelid closure. The definition of impaired joint position sensation was present if the examiner senses the correct sense under 1 out of 3. The definition of sensory ataxia was two or more of 1. Romberg's sign, 2. pseudoathetosis with impaired joint position and/or vibration sensation, and 3. absence of nystagmus and/or cerebellar dysarthria [4].

We examined 43 patients with impaired vibratory and/or joint position sensation (Table 1). The cause of symptoms and localization of anatomical lesions in patients with vibratory impairments were various. Most of patients (N = 35) developed sensory ataxia (with impaired vibratory sensation, N = 15; impaired joint position sensation, N = 3; both sensation impairments, N = 17) due to lesions in the nerve root/

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Table 1
Number and causes of patients with vibratory and/or joint position sensation impairments, and presence of sensory ataxia.

Localization	N	Cause of symptoms	Vibratory sensation impairment	Joint position sensation impairment	Both sensation impairments
Peripheral nerve	3/*1*	Sjögren's syndrome, POEMS syndrome, angiitis	2/0	0	1/*1*
Nerve root/dorsal root ganglion	22/*22*	Sjögren's syndrome, CIDP, POEMS syndrome, etc	11/*11*	0	11/*11*
Spinal cord	15/*12*	NMOsd, malignant lymphoma, cervical spondylotic myelopathy, syringomyelia, etc	6/*4*	3/*3*	6/*5*
Brainstem	3/*0*	medullary infarction (N = 2), pontine infarction (N = 1)	2/*0*	0	1/*0*
Cerebral lesion	0		0	0	0
Total	43/*35*		21/*15*	3/*3*	19/*17*

CIDP – chronic inflammatory demyelinating polyneuropathy; NMOsd – neuromyelitis optica spectrum disorder; POEMS – Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes.
Italic numbers present numbers of patients with sensory ataxia.

dorsal root ganglion or spinal cord. Interestingly, patients with sensory ataxia, who showed the dissociation between intact vibratory and impaired joint position sensation was observed only in the three spinal cord disorders in this cohort. Followings are case reports about the patients with intact vibratory and impaired joint position sensation.

2.1. Patient 1

An 85-year-old man without any history of trauma developed ataxic gait over 2 months. He needed a walker to walk. Neurological examinations showed limb and truncal ataxia, and severely impaired joint position sensation. However, his muscle strength, tendon reflexes and vibratory sensation were intact. In the thumb localization test, he showed ataxic movements. His nerve conduction velocity and amplitude in median, ulnar, tibial and sural nerves were normal. Central conduction delay (N13-N20) was observed in somatosensory evoked potentials. Brain MRI did not reveal cerebellar atrophy. Cervical MRI revealed a mass lesion at the retro-odontoid region without enhancement, which compressed the upper spinal cord (Fig. 1a). We diagnosed him as having a pseudotumor caused by atlantoaxial subluxation, after we ruled out primary or metastatic malignancy by systemic computerized tomography and gallium scintigraphy and inflammatory disorders such as rheumatoid arthritis or chronic kidney disease. The patient underwent C1–2 posterior spinal fusion. After the operation, his neurological signs and symptoms improved markedly and he was able to walk without any walking aids. In the thumb localization test, his ataxic movements also improved.

2.2. Patient 2

A 61-year-old man without any history of trauma was referred to our hospital for truncal ataxia that developed over a year. Neurological examinations showed limb and truncal ataxia, hyperreflexia in the lower extremities, and severely impaired joint position sensation. However, vibratory sensation was intact. He did not show any paresis. His nerve conduction velocity and amplitude in median, ulnar, tibial and sural nerves were normal. Brain MRI did not reveal apparent cerebellar atrophy. Cervical MRI revealed multi-level spinal cord compression caused by cervical spondylosis (Fig. 1b). We diagnosed him as having spinal ataxia due to cervical spondylotic myelopathy. The patient underwent C3–6 laminoplasty. After the operation, his neurological signs and symptoms improved markedly.

2.3. Patient 3

A 66-year-old man with flu episode was referred to our hospital for developing truncal ataxia that developed over several days. Neurological examinations showed limb and truncal ataxia, hyperreflexia in the extremities, and severely impaired joint position sensation. However, vibratory sensation was intact. He did not show any paresis. Brain MRI did not reveal apparent cerebellar atrophy. Spinal MRI revealed multi-level spinal cord hyperintensities in T2-weighted images (Fig. 1c). Anti-dsDNA, anti-SS-A, anti-SS-B, and anti-neutrophil cytoplasmic antibodies were not detected. Cerebrospinal fluid (CSF) showed 17

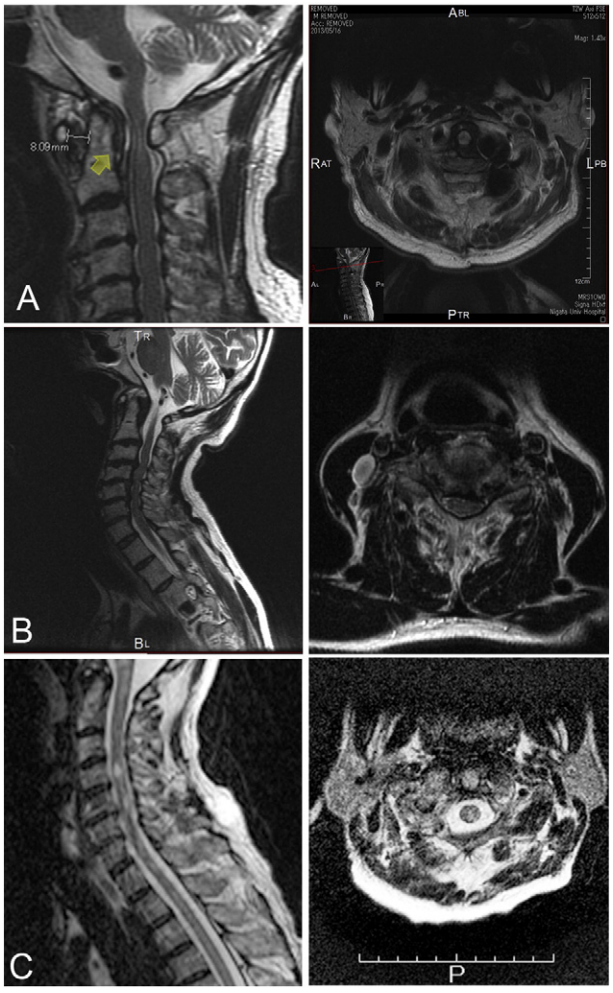

Fig. 1. Magnetic resonance images of cervical spinal cords. (A) Sagittal T2-weighted images (left) showing a mass lesion (arrow) at retro-odontoid region and upper cervical cord atrophy. The mass lesion showed no enhanced area (not shown). Sagittal T2-weighted images revealed a widening of atlantodental interval (8.09 mm). Arrow indicated pseudotumor. (B) Sagittal T2-weighted images (left) showing spinal cord compression at C3/4, C4/5, and C5/6 levels. On the axial T2-weighted image at the C4/5 level (right), spinal cord compression was prominent on the left side. (C) Sagittal T2-weighted images (left) showing spinal cord hyperintensites at C4 and C5 levels. On the axial T2-weighted image at the C4 level (right), spinal cord hyperintensity was localized in the posterior column.

leukocytes/mm^3. The CSF protein and glucose concentration was 84 mg/dL and 53 mg/dL. The level of myelin basic protein was 745 pg/mL. Negative polymerase chain reaction results for herpes simplex virus, varicella-zoster virus, Epstein-Barr virus, and cytomegalovirus were obtained in the CSF. We diagnosed him as having spinal ataxia due to acute disseminated encephalomyelitis. The patient was treated with prednisolone. Steroid therapy improved his neurological signs and symptoms markedly.

3. Discussion

Generally, vibratory sensation and joint position sensation are impaired simultaneously in patients with lesions of the dorsal columns. However, Netsky reported the case of a patient with syringomyelia in whom the vibration sensation was diminished and the joint position sensation was intact [2]. Pathologically, the dorsal column was intact, but demyelination was observed in the lateral funiculus. He inferred the presence of a pathway of vibratory sensation in the lateral funiculus. The ascending fibers for vibration sensation are likely to be divided into the dorsal column and lateral funiculus. Similar patients with impaired vibratory sensation and intact joint position sensation had been described in the patients with multiple sclerosis [3], rheumatoid cervical myelopathy [5], and brainstem infarction [6]. We did not find any reported patients with impaired vibratory sensation and intact joint position sensation who had developed spinal ataxia. To the contrary, we speculated that in our patients the dorsal column was mainly involved, but the lateral funiculus was spared because of less motor signs and symptoms.

The limitation of this study should be acknowledged. Because the sensitivities of impaired vibratory sensation identified by using a tuning fork and impaired joint position sensation by physical examinations are different, we would possibly underestimate the presence of impaired joint position sensation in this cohort. Additional studies are required to confirm our results.

4. Conclusion

We presented the cases of patients with retro-odontoid pseudotumor, cervical spondylotic myelopathy, and acute disseminated encephalomyelitis who developed spinal ataxia. We emphasize that the dissociation between the intact vibratory sensation and the impaired position sensation is an important sign of spinal ataxia even though patients did not show other myelopathic signs. The combination of intact vibratory sensation and impaired joint position sensation may predict ataxia of spinal origin.

Conflict of interest

The authors have no financial interest related to the material in the manuscript.

Authorship

(1) The conception and design of the study, or acquisition of data, or analysis and interpretation of data; MK, KK, TT, (2) drafting the article or revising it critically for important intellectual content; MK, NE, OO, MN, (3) final approval of the version to be submitted; MK, MN.

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